A CASE OF PRIMARY COMBINED COLUMN DISEASE.

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MRS. L , aged 59 years, presented herself for examination, complaining of unsteadiness in walking and numbness from the waist downward. Her family history reveals a decided neurotic taint. Her grandmother and three aunts had paralysis agitans, a sister nystagmus, and her mother an attack of apoplexy. She herself had enjoyed excellent health prior to the onset of the present affection, which began, she stated, suddenly, about two years previously.

There was an attack of unconsciousness preceded by stiffness in the neck and nausea. No paralysis followed, but shortly afterwards distinct insecurity in walking, with progressive weakness in both legs developed. She became emaciated and extremely pallid.

When examined by Dr. Frances Janney, eighteen months after the onset, she presented the following symptoms:

The patient was exceedingly anæmic, and moderately emaciated. There was no paralysis, though the legs were weak. The wasting was general and did not suggest atrophy from central trouble. The station was poor with feet together and eyes open or closed. The gait was distinctly ataxic. The knee jerks were decreased on both sides, but reinforcible. The plantar reflex was retained. There was no change in sensation except paræsthesia of the legs and trunk to the waist. A vaginal examination revealed the presence of a large tumor involving the fundus of the uterus. Its existence had hitherto not been suspected.

Some months later a second examination was made and the above condition confirmed. The knee jerks were irregular. On the right the response was slightly below normal, on the left almost normal, on both sides reinforcible. There was no clonus or ankle jerk. The elbow jerk and chin jerk were present. Muscle reaction was good. Pain sense and sensation to touch and heat were normal. The paræsthesia was still present.

There was no disturbance of the bladder or rectal functions. A fine rythmical tremor in both hands was observed. This had existed for years and was not made worse by voluntary effort. An examination of the blood made by Dr. James E. Talley, showed 2.464,000 red blood cells and 40% hæmoglobin. Dr. A. G. Thomson reported the eye condition as follows: Pupillary reaction normal, form fields and fundi normal. Clear media, margins of discs hazy. The patient died a year later from exhaustion. Her exact nervous condition at the time was not studied.

The necropsy revealed the following facts: In the uterus, involving the whole body of this organ, was a large, hard tumor, which microscopic section proved to be a fibro-sarcoma. Some few masses, small in size and scattered, were seen in the liver, which was normal in size. The examination otherwise proved negative, except the spinal cord, which was the seat of a lesion presenting the greatest interest. Microscopically the cord appeared smaller than normal. There was no meningitis. The brain and its enveloping membranes showed no change. Unfortunately, the brain in the process of hardening, underwent decomposition, hence a report of the microscopical condition of this organ is impossible. Macroscopically there was demonstrated degeneration in the posterior and lateral columns of the spinal cord, as seen by the distinct paling of these tracts. The process involved the whole extent of the cord and was most intense in the lower dorsal region. The degeneration in the lateral tracts was not so extensive as in the posterior columns at any of the levels.

The cord was hardened in Müller's fluid, imbedded in celloidin, and stained by the Weigert-Pal method and with ammonio-carmine.

Microscopical.—After staining, a sclerotic change was seen in the posterior and lateral pyramidal tracts throughout the whole extent of the cord, almost systemic in character. In the posterior columns the degeneration is most intense in the dorsal region, least in the lumbar region, while in the cervical region it is considerably more than moderate. There is always a strip of healthy tissue surrounding the degenerated area in the posterior columns

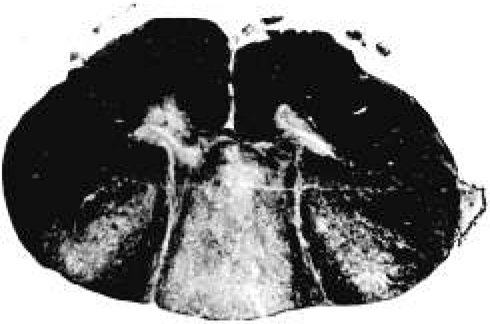

FIG. 1. Cervical cord showing degeneration of the postero-median, the postero-external (in part), and the crossed pyramidal tracts.

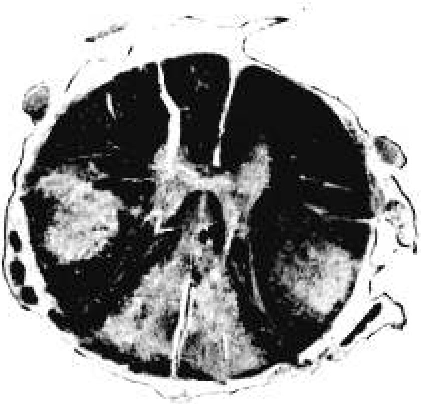

FIG. 2. Thoracic cord showing degeneration in the posterior and lateral columns.

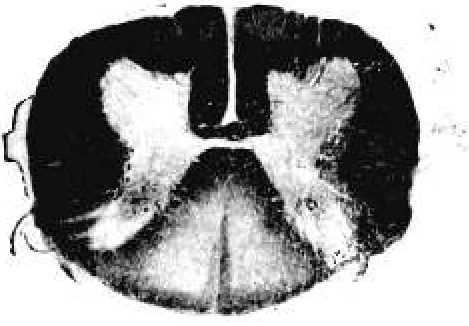

FIG. 3. Lumbar cord showing degeneration in the posterior columns.

and the columns of Goll are more intensely involved than the columns of Burdach.

The ganglion cells in the anterior horns show slight change in places, chiefly in the upper dorsal region. The posterior roots and Clark's column show no signs of involvement.

In the upper cervical region, the degenerated area involves the columns of Goll in their entirety, except a few fibres medianward and cervically, which take the stain well. Though to a much less degree, the columns of Burdach are markedly involved. Centrally there remain a number of healthy fibres, while peripherally but a few well-stained fibres can be seen. The root zones are present. In the lateral pyramidal tracts there is beginning degeneration, but to a much less degree than is seen in the posterior tracts. The anterior pyramidal and cerebellar tracts are intact. The anterior horns and posterior roots as well as the remaining tracts of the cord are normal.

In the cervical development, the same condition exists save that the degeneration in the postero-external columns and in the lateral pyramidal tracts is more intense.

In the dorsal region the process has so far progressed that few healthy fibres remain in Goll's columns. In the columns of Burdack a narrow strip along the posterior horns alone is stained, the process growing more intense from above downwards. In the anterior pyramidal tracts a very moderate degree of degeneration is observed. There is marked degeneration of the crossed pyramidal tract. In the upper levels a few ganglion cells have lost their prolongation, but there seems to be no lessening in the number. The process involved to a very slight degree the antero-lateral ascending tracts.

In the lumbar region the degeneration is less marked, especially marginally. In the posterior columns healthy fibres in increasing numbers are observed. A band of well-stained tissue encircles completely the degenerated area.

In the lumbar enlargement, only traces of the process in the lateral columns are seen. In the posterior columns are two wedge-shaped areas of degeneration on either side of the posterior fissure, surrounded by healthy tissue.

The degeneration now rapidly becomes less in extent and degree, until in the sacral cord there is no trace of the process visible. Clark's columns and the posterior roots are intact.

The carmine specimens show smaller axis-cylinders; in places many entirely disappeared; a marked increase in connective tissue. To a very moderate degree is there increase in the blood vessels. There were seen no granular cells in any of the sections.

This is evidently one of a group of cases which has been variously described as combined tabes, postero-lateral sclerosis, spastic tabes (Grasset), ataxo-paraplegic tabes (Déjerine), ataxic paraplegia (Gowers), combined fascicular disease, progressive spastic ataxia (Dana), true combined system disease (Babinski and Charrin) and primary combined column disease (Rothman).

In 1871 Prevost described the post-mortem findings in what was probably the first case belonging to this group. Later, Babesin, Strümpell, Siali, Westphal and others added cases to the literature of the subject. In 1886 Grasset collected 33 cases with autopsies and called the disease combined tabes (ataxo-spasmodique). The evidence did not point to a mere systemic lesion. He believed with Gowers that while the lesion in the posterior columns was systemic, in the lateral columns it tended to be diffuse, and concluded that the process was a diffuse myelitis.

A study of the autopsies in the cases above demonstrates clearly that the disease is one with a most varied morbid anatomy, different columns in varying degrees of intensity being involved. The gray matter may or may not be diseased; the peripheral nerves have been found involved; Clark's columns and the posterior roots as shown by Rothman, Mayer and others are often affected. On the other hand, a number of cases show a noteworthy unanimity in the distribution of the lesion in the posterior columns With moderate constancy the lesion is more intense in the lower dorsal region, less in the cervical region and least in the lumbar and sacral regions. As a rule there is a band of healthy fibres surrounding the diseased areas adjacent to the posterior horns. In the lateral columns the degeneration is not strictly systemic as Gowers, Dana and others have proven, and which is shown in the case under discussion.

In some cases the lateral mixed columns are encroached upon, and the direct cerebellar tracts are not infrequently involved. In Strümpell's case, reported in 1880, there was systemic degeneration of the pyramidal tracts, posterior columns, cerebellar tracts with atrophy of Clark's columns. The anterior horns and peripheral nerves escaped.

This variability in the morbid anatomy, together with the irregularity in the clinical history, suggests that the lesion is not truly systemic, but rather diffuse, beginning as a primary disease of the column of the cord, the posterior and lateral pyramidal tracts most usually, and developing as the process progresses into a diffuse one. It is not a systemic lesion, as Strümpell, Hochhaus and others maintained. It may, if the patient succumbs early, show a limitation to certain tracts in the cord, but when the disease is of long duration the process takes on an increasingly diffuse character. Strümpell believed that the further extension of the disease by continuity is improbable. Certainly, at first the limitation to certain columns, and the freedom of the parts immediately adjoining the diseased areas, inclines to this view, but later there is little doubt that continuity plays at least some part in the extension of the lesion.

The presence in the case under discussion, of a malignant tumor of the uterus is interesting in connection with the recent reports of spinal degeneration in constitutions with morbid blood states. Lichtheim, Nonne, Minnick, Burr and others have described characteristic lesions of the cord in pernicious anæmia. Babes and Kalindero, in 1890, reported a case of Addison's disease, with lesions in the posterior and lateral columns with a distribution which suggests to no slight degree that found in many of these cases of combined sclerosis. Fleiner quotes Abegg's case of spinal degeneration in a case of Addison's disease. There was change in the posterior columns in two of his own cases.

Minnich mentions spinal change in three cases of chronic icterus, in one case of leukæmia, in a case that died of tumor of the "vermis inf. cerebri," in two cases of chronic nephritis, and finally, a case of carcinomatous cachexia with hydræmia. Lastly, spinal lesion has been found in hypnotismus, pellagra, lathyrismus, and lead and alcohol poisoning. This clearly demonstrates some relation between toxic blood conditions and spinal degeneration, but just what that relation is, it is impossible in the present light of our knowledge to decide. In pernicious anæmia, the origin of the blood state remains as obscure as the cause of the spinal lesion. The theory that the two conditions are common results of the same cause, probably a toxine seems the most plausable explanation, one is justified at present in making.

Since the exact duration of the tumor of the uterus is not known, we are inclined to place no importance upon its presence, and all the more likely to consider its presence a coincidence.

In studying the etiology of the affection it will be seen that nervous heredity plays an unimportant part; that syphilis is rarely an antecedent; that it is more frequent in males; that it is a disease of adult life; that exposure and excesses, exercise, predispose, and lastly, it follows spinal injury according to Rothman. The symptoms are characteristics. The onset is gradual, the first symptom being unsteadiness in walking. Soon there is added stiffness in the muscles, though this may be absent. Romberg's symptom is present and the gait is distinctly ataxic, though differing from the walk of true tabes. The high raising and flopping down of the feet is not prominent. It is a mingling of the gaits of spastic paralysis and true tabes—when spastic symptoms are present. Lighting pains are almost always absent owing probably to the freedom of the root zones. Other sensory symptoms are rare. The reflexes show a wide difference from the tabes. The knee jerks are usually much exaggerated or spastic at first, while towards the last they may be almost or quite absent. The plantar reflex is present, as well as the remainder of the superficial reflexes. The sexual power is early lost. The eye symptoms sometimes, but rather exceptionally, resemble those found in true tabes. The iris reflex to light is usually preserved and optic atrophy is the exception. Progressive weakness ushers in the final symptoms.

The disease has little tendency in itself to cause death according to Gowers and Dana, and usually is of long duration. Rothman's experience, however, leads him to conclude that the duration of the disease is but three years. In our own case the patient expired just three years after the appearance of the first symptoms, but the case was not uncomplicated, and had the malignant tumor been absent we are warranted in supposing that the patient would have survived a longer period.

In conclusion, thanks are due Dr. Frances Janney for her kindness in permitting this report and for the opportunity of making the necropsy. To the kindness of Dr. Schively are we indebted for the photographs.

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